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Case Report

A late presentation of an anomalous left coronary artery originating from the pulmonary artery (ALCAPA): A case study and review of the literature

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ABSTRACT

Background: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), also known as Bland–White–Garland syndrome, is a rare congenital condition which can manifest as various cardiac symptoms.
Case report: A 66-year-old woman who presented for pre-operative surgical risk assessment for hip surgery underwent a nuclear stress test which revealed a large reversible anterior defect. At coronary angiography she was found to have ALCAPA.
Conclusions: This is a rare case of ALCAPA due to the patient's age. Survival to adulthood is possible and patients may remain relatively asymptomatic for years.
<**Learning objective:** Discuss the common clinical manifestations of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). Discuss the imaging findings of ALCAPA. Discuss the treatment options for ALCAPA.>

Introduction

The anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA, also known as Bland–White–Garland syndrome) is a rare congenital coronary artery anomaly, occurring in 0.26% of the patients with congenital heart disease [1]. Common clinical manifestations include dilated cardiomyopathy, dyspnea, angina pectoris, signs of ischemia on the electrocardiogram, heart failure, sudden death, and death in childhood [2]. Up to 90% of the patients with this condition die during the first year of life because of left ventricular failure [1]. ALCAPA should be considered when the differential diagnosis includes dilated cardiomyopathy, coronary artery fistula, mitral regurgitation, and myocarditis.

Case report

A 66-year-old woman with a history of chronic obstructive pulmonary disease (COPD), obstructive sleep apnea (OSA), pulmonary hypertension, inflammatory bowel disease, arthritis with degenerative joint disease, right hip pain with multiple falls, which confined her to a wheelchair for the previous 6 months, presented for a preoperative cardiovascular risk assessment for hip surgery. The patient's last cardiovascular evaluation was 7 years prior due to complaints of shortness of breath (SOB), which was attributed to pulmonary hypertension from underlying COPD and OSA. Her review of systems was positive for SOB with moderate activity, although she reported no recent SOB as her activity has been limited to her hip pain, chronic lower extremity edema, a remote history of paroxysmal nocturnal dyspnea and chest pain, occasional palpitations, but no syncope, and multiple falls, which had confined her to a wheelchair. Current medications included: digoxin, furosemide, spironolactone, mesalamine, albuterol, and fluticasone. She had a family history of coronary artery disease and denied any history of smoking. On examination, she was hemodynamically stable with a heart rate of 86 beats per minute (bpm), blood pressure of 126/68 mmHg, respiratory rate of 18, and pulse oximetry of 95% on room air. Pertinent physical examination findings included chronic bilateral 2^{+} pitting ankle edema and the absence of a cardiac murmur or pulmonary rales. Seven years previously an echocardiogram reportedly showed a normal left ventricular ejection fraction (LVEF) and normal valvular function. As part of her current preoperative risk assessment the patient

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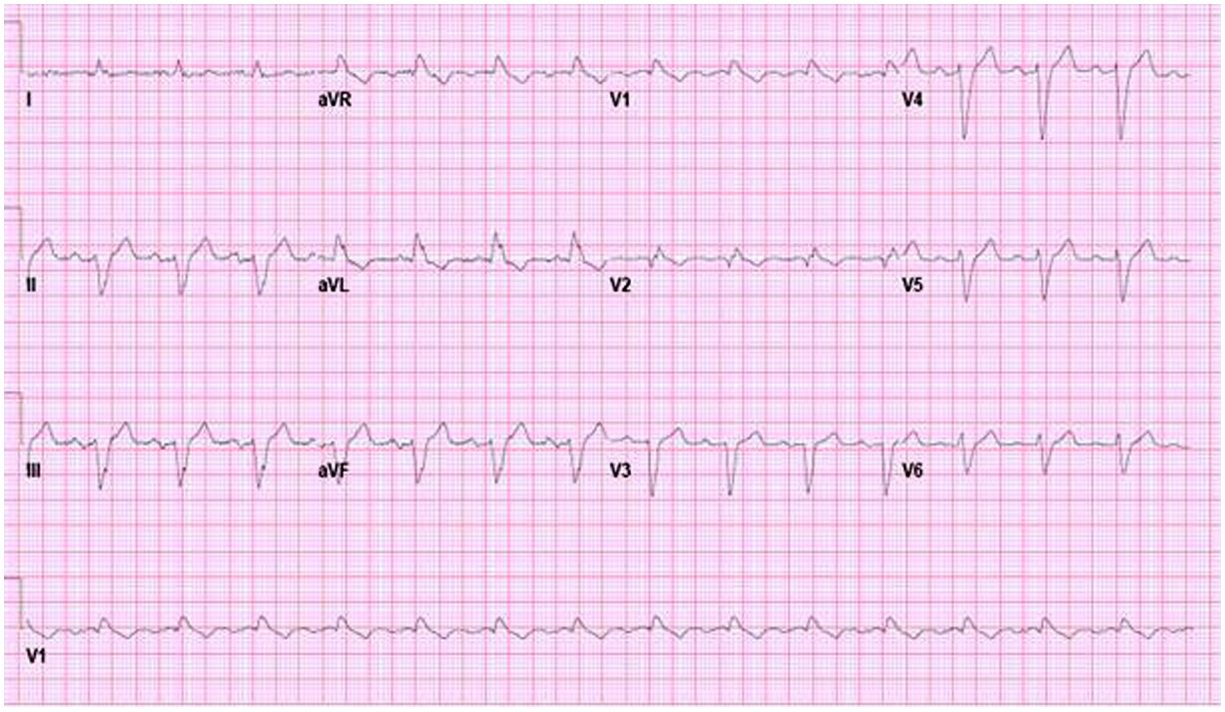

Fig. 1. Electrocardiogram: sinus rhythm with left-axis deviation, right bundle branch block, and anteroseptal infarct; age undetermined.

underwent an electrocardiogram (ECG) and dipyrimadole nuclear stress test. The ECG did not show any signs of ischemia (Fig. 1). The dipyrimadole/*technetium sestamibi* stress test revealed a large reversible anterior, anteroseptal, inferolateral, and apical defect, with an LVEF of 31%. A repeat echocardiogram was not performed despite the finding of a newly depressed LVEF.

Due to the potentially reversible changes seen on the nuclear scan and symptoms, the patient underwent cardiac catheterization. Hemodynamically, her cardiac catheterization showed no systemic hypertension, mildly elevated left ventricular end diastolic pressure (20 mmHg), mildly elevated pulmonary capillary wedge pressure (17 mmHg), normal systemic vascular resistance (1006 dyn/s/cm^2), normal pulmonary vascular resistance (267 dyn/s/cm^2), and moderate pulmonary hypertension (pulmonary arterial pressure of 62/29 mmHg, with a mean of 41 mmHg). Her global left ventricular function was moderately depressed and her LVEF calculated by contrast ventriculography was 35%.

While evaluating her vasculature she was noted to have a dilated pulmonary artery with the left main coronary artery originating posteriorly on the pulmonary artery angiogram (Fig. 2). Her left main trunk was moderately tortuous and moderately ectatic. Its distal portion was supplied by extensive collaterals from the right coronary artery (RCA). The RCA was large sized (dominant) and moderately ectatic (Fig. 3). Aortography showed

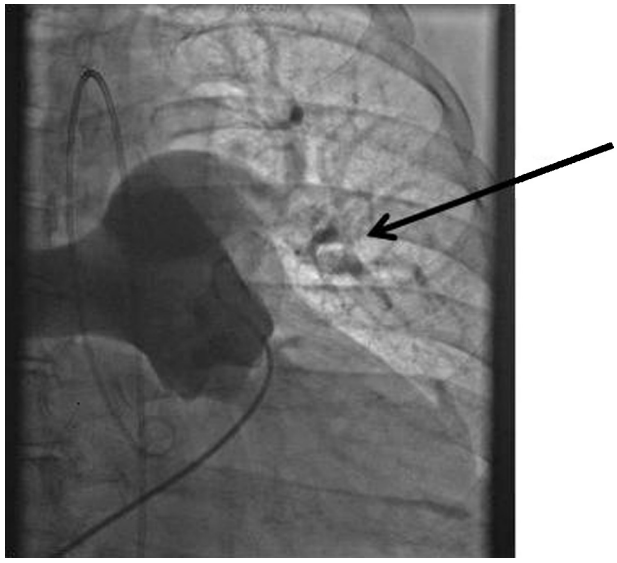

Fig. 2. Pulmonary artery angiogram showing the left main coronary artery arising from the left pulmonary artery.

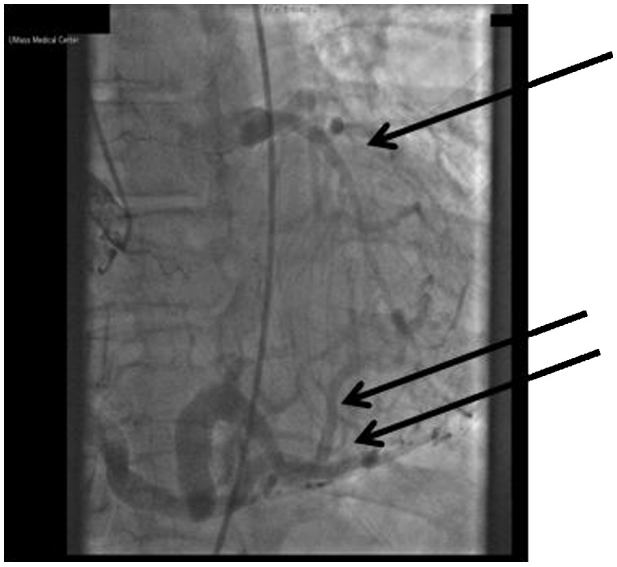

Fig. 3. Right coronary artery (double arrow) with tortuosity and collateralization to the left coronary artery (single arrow).

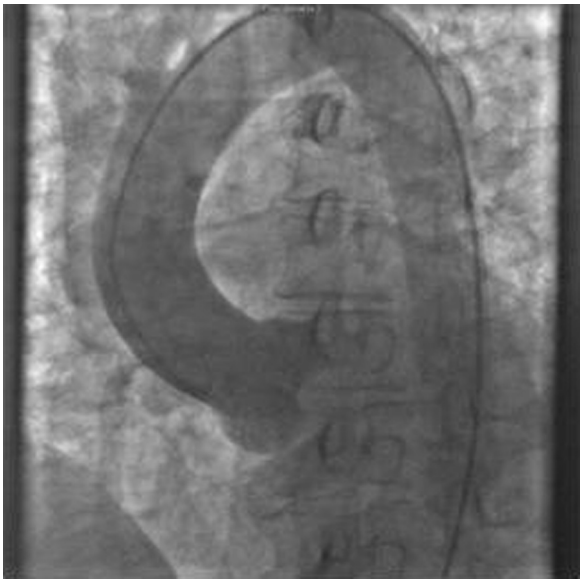

Fig. 4. Aortic root shot (absence of the left coronary ostium).

no evidence of disease. The aortic root and ascending aorta were satisfactorily visualized (Fig. 4). Overall impression was no significant coronary artery disease and an ALCAPA.

Cardiac surgery was consulted, and it was determined that the patient could undergo her hip surgery from a cardiac standpoint. It was determined that she would require further cardiac workup to define her anatomy with magnetic resonance imaging (MRI)/magnetic resonance angiography along with a viability study. She underwent successful, uneventful hip surgery on medical therapy. Unfortunately, following the surgery, the patient moved out of the region, so additional follow-up, including imaging, could not be performed.

Discussion

ALCAPA was first described in 1911 by Abrikossoff, a Russian pathologist, while reporting the autopsy of a 5-month-old infant [3]. In 1933, 22 years later, an article by Bland et al. appeared in the *American Heart Journal*, entitled "Congenital anomalies of the coronary arteries: report of an unusual case associated with cardiac hypertrophy" in which he described a 3-month-old boy with grunting, pallor, and cold sweats during nursing. Physical examination failed to demonstrate an appreciable murmur or cyanosis, but was remarkable for an increased area of cardiac dullness. Unfortunately the boy died at 3.5 months. The discovery of the origin of the left coronary artery arising from the pulmonary artery was made during the autopsy [4].

ALCAPA has an estimated incidence of 1/300,000 births [5]. This estimate may not accurately reflect the true incidence, as many patients may be asymptomatic until their death and therefore remain undiagnosed [6]. Most signs and symptoms are undetectable at birth because blood pressure in the pulmonary arteries is still similar to the systemic circulation. Only after a few months, as blood pressure falls in the pulmonary circuit, do symptoms become unmasked. The development of collateral circulation between the right and left coronary arteries determines the extent of myocardial ischemia; thus, patients with well-established collaterals are classified as the "adult type", and those with no collaterals as the "infantile type" [7]. For those individuals with the "adult type" of ALCAPA, pulmonary hypertension gradually develops. Pulmonary hypertension occurs because of left-to-right shunting as blood passes in a retrograde fashion from the right coronary artery to the left coronary artery by collateral circulation, and finally exiting the left main coronary artery into the pulmonary artery.

Patients with ALCAPA who survive past childhood often have varying symptoms of myocardial ischemia or heart failure, depending on the development of collateral circulation. Sudden cardiac death occurs in up to 90% of the patients at a mean age of 35 years [8]. Patients rarely survive to the sixth or seventh decade of life [9]. In 1995, Alexi-Meskishvili and colleagues found only 26 patients over 50 years old [10]. These figures confirm that our relatively asymptomatic 66-year-old patient is a noteworthy case.

The diagnosis of ALCAPA is often made during coronary angiography. Angiography will demonstrate a dilated tortuous right coronary artery with collateral filling of the left coronary system. There are also noninvasive studies, which may provide key features to point the physician toward this coronary anomaly. An ECG may demonstrate left-axis deviation, left ventricular hypertrophy, poor R wave progression, and abnormal Q-waves in leads I and aVL [7]. Echocardiographic evidence includes a dilated right coronary artery and continuous blood flow from the left coronary artery into the pulmonary artery and no obvious origin of the left main coronary artery from the aortic root [11]. Additional findings include increased echogenicity of papillary muscles, and visualization of the left main coronary artery from the pulmonary trunk [11]. Additional imaging studies such as cardiac MRI with delayed gadolinium enhancement and cardiac computed tomography (CT) angiography have been performed to make the diagnosis of ALCAPA [12]. MRI and CT can provide direct visualization of the left coronary artery arising from the pulmonary artery. However, MRI can also show reversal of flow from the left coronary artery into the pulmonary artery which can be visualized with cine imaging [13].

Several treatment options are available with the primary objective of stopping blood escape from the coronary circulation to low pressure pulmonary circulation [14]. The first technique involved the ligation of the left coronary artery due to collateral circulation from the right coronary artery. Other options include coronary artery bypass grafting or surgical reconstruction creating a two coronary artery system [15]. The goals of current surgical management include establishing a two-coronary system with long-term patency, use of native tissue, and maintenance of potential for normal growth of coronary ostia and arteries [15].

Our patient, aged 66 years, was incidentally discovered to have ALCAPA during a cardiac catheterization. Another interesting feature, other than her age, is her previous diagnosis of pulmonary hypertension. This diagnosis had previously been attributed to the effects of underlying COPD and OSA. However, as seen during her cardiac catheterization, pulmonary hypertension may have resulted from her previously undiagnosed coronary artery anomaly.

Conclusions

The ALCAPA is a rare congenital disorder. Our 66-year-old patient had a history of pulmonary hypertension, but no chest pain to suggest myocardial ischemia. Her coronary artery anomaly was only diagnosed after invasive studies as part of a pre-operative risk assessment for elective hip surgery. Her case is significant because of her age and her features that are consistent with the "adult type" of ALCAPA, which demonstrate that an extended adult survival is possible and a patient may remain relatively asymptomatic for years.

Conflict of interest

The authors declare no conflict of interest.

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